Pheochromocytoma manifesting with shock presents a clinical paradox: a case report

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Abstract

A 46-YEAR-OLD MAN PRESENTED WITH SHOCK and adult respiratory distress syndrome. Investigations revealed an adrenal mass that was diagnosed, by fine-needle aspiration biopsy, as pheochromocytoma. Because biopsy is contraindicated in patients with pheochromocytoma, this confusing presentation underscores the value of excluding this diagnosis by biochemical means before performing fine-needle aspiration of adrenal tumours.

Résumé

UN HOMME DE 46 ANS S’EST PRÉSENTÉ EN ÉTAT DE CHOC et d’insuffisance respiratoire aiguë des états de choc. Les examens ont révélé une masse surrénalienne qu’on a diagnostiquée comme un phéochromocytome après avoir procédé à une biopsie par aspiration à l'aiguille fine. Comme la biopsie est contre-indiquée chez les patients atteints d’un phéochromocytome, toute présentation qui suscite la confusion démontre qu’il vaut la peine d'exclure ce diagnostic par des moyens biochimiques avant de procéder à une biopsie d’une tumeur surrénalienne par aspiration à l’aiguille fine.

Pheochromocytoma is an uncommon catecholamine-producing neoplasm, which usually presents with hypertension and the symptom triad of headaches, diaphoresis and palpitations.1–3 Clinical awareness of the protean manifestations, both typical and atypical, of pheochromocytoma is key to appropriate management.4 When pheochromocytoma presents with hypotension, clinicians may not include this tumour in the differential diagnosis and may expose the patient to unnecessary and potentially life-threatening risks during investigations.5 We describe a patient presenting with shock and adult respiratory distress syndrome in whom the initial working diagnosis was septic shock. During the work-up, ultrasonography revealed an adrenal mass. Fine-needle biopsy indicated pheochromocytoma. After the biopsy procedure, the patient experienced symptoms similar to those recorded at the onset of his illness.

Case report

A 46-year-old man arrived at a small community hospital with nausea, vomiting and abdominal pain of 1 day’s duration. He gave a recent history of borderline diabetes controlled by diet and a remote history of peptic ulcer disease. The patient’s temperature was 35.9°C, blood pressure 80/64 mm Hg, pulse rate 142 beats/minute and respiratory rate 36 breaths/minute. There was bilateral tenderness of the costovertebral angle.

The hemoglobin level was 160 g/L and the leukocyte count 21 × 10⁹/L, with neutrophilia. The level of urea nitrogen was 10.9 mmol/L and that of creatinine 257 µmol/L. Urine dipstick tests indicated 0.3 g/L protein and 250 erythrocytes/L. Microscopic urinalysis revealed 6 to 15 leukocytes and 11 to 20 erythrocytes per high-power field and 1 to 3 granular casts per low-power field. The level of total creatine kinase (CK) was 5716 (normally 0–243) U/L and that of the...
After administration of 1.5 L Ringer’s lactate solution, the patient was given 2 g cefotaxime, 500 mg hydrocortisone, 180 mg furosemide and 50 mg ranitidine by intravenous bolus, oxygen by mask and up to 7.5 μg·min⁻¹·kg⁻¹ dobutamine by intravenous infusion. He was then transferred to a regional facility.

On arrival, the patient was alert, oriented and diaphoretic; his skin was grey, and there was peripheral cyanosis. Skin turgor was normal. The temperature was 38.4°C, blood pressure 70/60 mm Hg, pulse rate 160 beats/minute (weak) and respiratory rate 48 breaths/minute. Arterial blood gas analysis of samples obtained while the patient was receiving 100% oxygen gave the following results: pH 7.33, partial pressure of carbon dioxide 37 mm Hg, partial pressure of oxygen (Pₒ₂) 46 mm Hg by mask and up to 7.5 μg·min⁻¹·kg⁻¹ dobutamine by intravenous infusion. He was then transferred to a regional facility.

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Erythromycin (500 mg every 6 hours) and gentamicin (400 mg daily), administered intravenously, were added to the antibiotic regimen. The therapy was adjusted until the patient eventually became afebrile and was hemodynamically stable. During the first 3 days in the unit, the patient was hypotensive, and an infusion of dobutamine supplemented with dopamine was needed to keep the mean arterial blood pressure above 65 mm Hg. On day 3, a right adrenal mass measuring 5 × 4 cm was detected by abdominal ultrasonography. Between days 3 and 6, there were 2 brief (lasting less than 1 hour) hypertensive episodes, both of which resolved after sublingual administration of 20 mg nifedipine. After day 6, the blood pressure was normal without further administration of antihypertensive agents. On day 7, adrenocorticotropic hormone stimulation yielded a peak response of 873 nmol/L, which indicated that adrenocortical function was normal. On day 10, the patient was extubated, and Pₒ₂ remained at 71 mm Hg with the patient receiving 40% oxygen. The patient was then transferred to a general medicine ward.

On day 14 fine-needle biopsy of the right adrenal mass was performed and pheochromocytoma diagnosed. Eight hours after the procedure, the patient vomited, became sweaty, and complained of shortness of breath and tightness of the chest. He volunteered that his symptoms were identical with those he had experienced at the onset of his illness. The blood pressure reached 240/120 mm Hg, pulse rate 100 beats/minute and respiratory rate 30 breaths/minute. The blood pressure dropped to 205/100 mm Hg after treatment with oxygen and nitroglycerin (administered transdermally). Electrocardiography at that time showed sinus tachycardia, and the level of CKMB was slightly elevated. Nine hours after the onset of this episode, the blood pressure was 90/60 mm Hg. Fig. 1 shows the changes in blood pressure after biopsy. Three days later, 24-hour urine catecholamine studies revealed a normal norepinephrine level of 442 (normally 70–500) nmol/d and a grossly elevated epinephrine level of 1126 (normally 10–130) nmol/d.

On day 28, after preparation with α- and β-adrenergic blockers and infusions of saline, right adrenalectomy was performed. The recovery was uneventful, and the patient was discharged from hospital 10 days after the surgery.

**Comments**

At presentation this man had an illness that was neither paroxysmal nor hypertensive, a combination that made the diagnosis of pheochromocytoma extraordinarily improbable.⁴,⁷ Instead, the patient was in shock, needing hemodynamic and respiratory support. Twice during his treatment for presumed sepsis, a known hypodynamic condition,⁷ he displayed transitory hypertension. Because the hypertensive episodes necessitated minimal therapeutic intervention, the care providers failed to appreciate their unusual significance.

Hypotension is the cardinal manifestation associated with epinephrine-secreting pheochromocytomas.⁵,⁶ Some authors have reported paroxysms of hypotension preceded by hypertension, similar to this patient’s post-biopsy attack.⁴,⁷ Although shock, pulmonary edema and tachycardia have been described in association with epinephrine-secreting pheochromocytomas,⁷⁶,⁷⁷ the presentation is rare enough to be absent from standard texts.⁷⁸,⁷⁹ We pos-
tulate that catecholamine-induced cardiac dysfunction combined with rapid administration of fluid led to pulmonary edema during the patient’s initial episode. The borderline CKMB levels associated with both attacks support this premise. This patient differs from those described in previous reports in that he had no history of attacks and there was no apparent trigger for the presenting episode.

Epinephrine is a catecholamine with greater affinity for the α-adrenergic than the β-adrenergic receptors. At low doses, epinephrine produces tachycardia via β-receptors in the heart and hypotension via α-receptors in the blood vessels of skeletal muscles. High doses of epinephrine produce hypertension via α-receptors in the peripheral vascular bed. This phenomenon may underlie the intraoperative hypertensive crises associated with epinephrine-secreting pheochromocytomas. The manipulation of the neoplastic gland may induce a burst of epinephrine secretion, which in turn leads to powerful α-mediated vasoconstriction that overwhelms the hypertensive effects of the β-mediated vasodilation.

In a recent review, Cook and Loriaux stated that hormone screening for adrenal tumours should be “tailor made” for the clinical context. The authors suggested that the clinical findings are a reliable guide for the selection of cases for biochemical testing. The diagnostic algorithm presented in the review calls for fine-needle aspiration biopsy of hormonally inactive masses. Pheochromocytoma is the one adrenal tumour that carries a higher risk for complications of aspiration. The possible complications are hemorrhage, because of the highly vascular nature of the tumour, and hypertensive crises due to catecholamine release. If complications occur, they may increase morbidity and result in death. The authors suggested that catecholamine screening for adrenal tumours should be “tailor made” for the clinical context. The authors suggested that the clinical findings are a reliable guide for the selection of cases for biochemical testing. The diagnostic algorithm presented in the review calls for fine-needle aspiration biopsy of hormonally inactive masses. Pheochromocytoma is the one adrenal tumour that carries a higher risk for complications of aspiration. The possible complications are hemorrhage, because of the highly vascular nature of the tumour, and hypertensive crises due to catecholamine release. If complications occur, they may increase morbidity and result in death.